

Anaesthesia for Children with Skin Disorders

Dr Catherine Riley^{1†}, Dr Kate Wilson²

¹ST7 Anaesthesia, Sheffield Children's Hospital, Sheffield, UK

²Consultant Anaesthetist, Sheffield Children's Hospital, Sheffield, UK

Edited by: Dr Faye M. Evans, Senior Associate in Perioperative Anaesthesia, Boston Children's Hospital, Boston, MA, USA; Dr Yuanting Zha, Acting Assistant Professor, Department of Anesthesiology, Seattle Children's Hospital, University of Washington, Seattle, WA, USA

†Corresponding author email: catherine.riley9@nhs.net

Published 27 August 2024

DOI: 10.28923/atotw.530



KEY POINTS

- Skin disorders are common in childhood, and, while most are mild, they can be life threatening.
- Skin disease can impact every stage of the anaesthetic and should be evaluated as part of the preoperative assessment.
- Treatments for skin disorders can also impact anaesthetic care.
- Anaesthetic challenges in these patients are many. The anaesthetist must balance prevention of further skin trauma or infection with the safe provision of anaesthesia.
- A multidisciplinary approach is recommended for severe cases, which should include the involvement of a dermatologist and/or treatment in a specialist centre.

INTRODUCTION

The skin is the largest organ of the body, and skin disorders are a common complaint of childhood. Skin disease may be the primary condition or the secondary manifestation of an underlying systemic disorder or genetic syndrome (Table 1). The severity of skin disease can vary from minor to severe and life-threatening. Children can present for general anaesthesia with coincidental skin disease or it may be the reason for their presentation.

Skin has several functions, including immunological and physical protection of the underlying organs, thermoregulation, and regulation of fluid balance. The anaesthetic implications of many skin disorders are manifestations of these functions and are broadly similar between individual dermatological diagnoses. The following are specific areas of concern:

- Airway
- Fluid balance
- Thermoregulation
- Infection risk
- Difficult intravenous access
- Difficulty securing lines, tubes, monitoring, or dressings

This tutorial will discuss some of the spectrum of skin disorders that a child may present with and the factors the anaesthetist may need to consider.

An online test is available for self-directed continuous medical education (CME). It is estimated to take 1 hour to complete. Please record time spent and report this to your accrediting body if you wish to claim CME points. A certificate will be awarded upon passing the test. Please refer to the accreditation policy [here](#).

[TAKE ONLINE TEST](#)

Subscribe to ATOTW tutorials by visiting <https://resources.wfsahq.org/anaesthesia-tutorial-of-the-week/>

Category	Condition	Dermatological Manifestations	Anaesthetic Implications
Vasculitic disorders	Behçet's disease	Intraoral and genital ulcers	<p>Friable intraoral ulcers may lead to:</p> <ul style="list-style-type: none"> • trauma • oropharyngeal scarring leading to difficult intubation <p>Systemic steroid therapy (see Table 2)</p> <p>Risk of difficult mask ventilation from facial asymmetry</p> <p>Risk of bleeding from facial angiomas</p> <p>Avoid nasal instrumentation</p> <p>Risk of bleeding from airway instrumentation</p>
	Sturge-Weber	Facial capillary angiomas (port-wine birthmark)	
	Osler-Weber-Rendu syndrome (hereditary haemorrhagic telangiectasia)	Telangiectasias occur on the skin, nasal mucosa (causing epistaxis), and gastrointestinal tract (causing gastrointestinal bleeding)	
Collagen/elastin disorders	Cutis laxa	Loose and inelastic skin and joints	<p>Careful positioning required to prevent joint dislocation</p> <p>Difficulty securing lines and tubes</p> <p>Difficult venous access</p> <p>Increased risk of bleeding and bruising; ensure adequate padding underneath the blood pressure cuff</p> <p>Difficulty securing lines and tubes</p>
	Ehler-Danlos	Skin elasticity and skin and vascular fragility	<p>Difficult airway management due to microstomia</p> <p>Systemic steroid use (see Table 2)</p> <p>Difficult intravenous access due to skin thickening</p>
Autoimmune skin disorders	Scleroderma	Skin and mucosal telangiectasias and skin thickening	<p>Systemic steroid use (see Table 2)</p> <p>Difficult airway management from intraoral blisters and scarring of oropharyngeal tissue</p> <p>Difficult venous access</p> <p>Care must be taken when securing tubes and lines; avoid adhesives</p>
	Bullous pemphigoid	A rare autoimmune bullous skin disease with widespread, tense bullae	<p>Edentulous patients may be difficult to mask ventilate</p> <p>Anatomical differences such as hypoplastic maxilla may make airway management difficult</p> <p>Lack of sweat glands can lead to hyperthermia</p> <p>Continuous temperature measurement and active cooling may be needed</p>
Disorders involving sweat glands/hair	Christ-Siemens-Touraine syndrome	An X-linked recessive disorder characterised by a lack of sebaceous or sweat glands and absent hair and teeth	

Category	Condition	Dermatological Manifestations	Anaesthetic Implications
Exfoliating skin conditions	Hypohidrotic ectodermal dysplasia	A rare syndrome characterised by abnormal ectodermal tissues: abnormal dentition, sparse hair, decreased sweating, and facial abnormalities	Lack of sweat glands can lead to hyperthermia Continuous temperature measurement and active cooling may be needed
	Stevens-Johnson syndrome and toxic epidermal necrolysis	A potentially fatal skin reaction to medication or infection characterised by fever and malaise and develops into blistering and desquamation of skin and mucous membranes	Difficult airway management from mucous membrane desquamation Need for strict asepsis Difficult venous access and securing lines and tubes Need for careful handling when moving to operating table and positioning for surgery to prevent further skin damage Systemic steroid use (See Table 2)
	Ritters disease (pemphigus neonatorum or staphylococcal scalded skin syndrome)	A disorder of infants caused by staphylococcal exotoxins, which break down desmosomes and cause widespread blisters	Risk of fluid loss Impaired thermoregulation and risk of hypothermia Risk of secondary infection

Table 1. Dermatological manifestations of the congenital syndromes and diseases associated with skin disorders

EXAMPLE 1

A 6-year-old is booked for urgent laparoscopic appendicectomy. Past medical history includes eczema treated with a paraffin-based emollient cream and topical steroid cream.

Dermatitis

Eczema (or atopic dermatitis) is common; 20% of children will experience eczema during their lifetime.¹ It is characterised by itchy, dry skin. The severity varies, but even mild cases may be treated with ointments, which can impair the adhesion of dressings used to secure an endotracheal tube or intravenous cannula. Paraffin-based creams, commonly used for treatment, make it difficult to attach electrocardiograph (ECG) electrodes. More severe cases may be treated with topical or systemic steroids. Children with atopic dermatitis are more likely to be diagnosed with asthma.²

Another common condition is contact dermatitis caused by chemical or physical irritants in contact with the skin. This may include “nappy rash” around the genitals or “dribble rash” around the mouth and chin. Treatment is usually with emollient or barrier cream, which can impair adhesion of tapes used to secure the endotracheal tube. Zinc oxide, an ingredient in some barrier creams, can lead to burns in a magnetic resonance imaging scanner.³ It may also be prudent to avoid local anaesthetic cream in children prone to contact dermatitis to avoid triggering a flare.⁴

EXAMPLE 2

A 10-year-old with epidermolysis bullosa (EB) is listed for elective syndactyly repair.

Epidermolysis Bullosa (EB)

This term describes a group of disorders characterised by bullae (blisters). The pathology is at the epithelial basement membrane zone, which allows formation of blisters after only minimal traction or trauma. There are 3 main types⁵:

- EB simplex: intradermal blisters (blisters form above the basement membrane); usually confined to hands and feet.
- Junctional EB: blisters form within the basement membrane, commonly the gastrointestinal tract and larynx; often fatal.
- Dystrophic EB: blisters develop below the basement membrane.

There are 2 genetic inheritance patterns, dominant and recessive; the recessive variant is more severe. The disease is characterised by vesicles and bullae, which can occur spontaneously or from minimal friction or trauma. When the vesicles and bullae heal, scarring can occur, leading to contractures and syndactyly. Patients can suffer from both acute and chronic pain. Treatment is supportive.⁵

Dilated cardiomyopathy is a serious complication of EB. Patients with severe recessive dystrophic EB are most at risk of dilated cardiomyopathy or congestive heart failure.⁶

The anaesthetist must balance prevention of new bullae and management of previously scarred tissue. The child’s airway can pose a challenge if such scarring causes microstomia or if there are active bullae present.

Special consideration should be given to the surgical setting. An elective operation should be undertaken in a specialist setting with input from the multidisciplinary EB team, which includes a consultant dermatologist, EB specialist nurse, and theatre and nursing staff familiar with the care of children with EB.⁷ Advantages of this setting include staff familiarity with skin precautions and the rapport that the patient may have with the specialist nurse who can help facilitate smooth induction to avoid skin trauma. Surgeons in specialist centres will have experience in treating the surgical complications of severe skin disease. However, children may present as emergencies to their local hospital, so all anaesthetists should be familiar with the principles of anaesthesia for children with these diseases. Communication between theatre and ward staff is vital in the nonspecialist centre, with clear plans disseminated throughout the perioperative period. Advice may still be sought from the specialist centre to guide treatment.

EXAMPLE 3

A baby with congenital ichthyosis requires release of skin constriction causing digital ischemia.

Ichthyosis is a term used to describe skin that continually scales, becoming very dry and thick. There are over 20 conditions that are characterised by ichthyosis, which can be congenital or acquired.⁸ The severity ranges from mild to life-threatening depending on the condition.

Ichthyosis vulgaris is an autosomal dominant common condition affecting 1 in 250 people and causes fine scaling of the limbs; it is usually mild and treated with emollients. More severe are the following autosomal recessive congenital ichthyosis disorders.

Harlequin-type ichthyosis is the most severe of these disorders. It is rare, with an incidence of 1 in 300000 live births.⁹ It causes thickened dry skin over the entire body. Admission to neonatal intensive care is required from birth. There is high

neonatal mortality due to sepsis and respiratory failure, secondary to impaired ventilation from restricted chest excursion. Thermoregulation is impaired secondary to large volumes of fluid loss, and the hyperkeratotic skin can cause digital or limb ischaemia through constriction. Treatment is largely supportive with regular skin debridement.

Congenital ichthyosiform erythroderma occurs in 1 in 300000 births and is the most common underlying cause of “collodion baby”, a baby born with a collodion membrane, a tight yellow film over the skin.¹⁰ The baby requires nursing in a neonatal critical care facility until the membrane is shed after about 1 week. In 10% of cases, the underlying skin is normal (“self-healing collodion baby”). Treatment is largely supportive and aims to prevent the complications of fissures that can form if the collodion membrane dries and cracks. Fissures can result in infection, temperature dysregulation, and fluid loss. The skin is shed over the first few weeks of life. The collodion membrane should not be debrided.

Lamellar ichthyosis occurs in 1 in 200000 births and presents similarly with a collodion membrane, progressing to hyperkeratosis on the palms, soles, and joints such as axillae, elbows, and neck.⁸

In both the above conditions, the underlying skin is red and becomes scaly with alopecia, ectropion, and the potential for digital ischaemia. Sweating is impaired, causing disordered temperature regulation and risking hyperthermia during exercise or in hot climates. Treatment is with emollients, prevention of hyperthermia, eye drops if ectropion is present, and, in severe cases, systemic retinoids.¹⁰

MEDICATION FOR SKIN DISEASE

Children with skin disease may be taking medication with implications for anaesthesia (Table 2).¹¹ Systemic exogenous steroids may cause adrenal suppression and therefore have major implications for anaesthesia. Oral retinoids, which are teratogenic, may be taken as treatment for acne vulgaris. The British National Formulary for Children advises effective contraception for females of child-bearing age during treatment with retinoids and for at least 1 month after cessation.¹¹ Such patients should be counselled to use alternative contraception if sugammadex is used during anaesthesia, as it can decrease the efficacy of oral contraceptives for up to 7 days.¹¹

PREOPERATIVE CONSIDERATIONS

History

Ask specifically about the severity of the skin disease, active infections, systemic involvement (such as an associated syndrome), or complications. Take a detailed drug history, including topical treatments. Children with EB may have an oesophageal stricture that

Drug Class	Example Drug Names	Indication	Mechanism of Action	Side Effects	Implications
Retinoids	Oral: Isotretinoin Acitretin Alitretinoin	Ichthyosis Acne vulgaris	Regulate gene transcription and aim to normalise keratinocyte proliferation	Anaemia Deranged liver function Rhabdomyolysis Teratogenic	Consider checking relevant laboratory studies Patients taking oral retinoids may also be taking oral contraceptives. If sugammadex is used, appropriate counselling should be given as it impairs the efficacy of oral contraceptives Lubricate eyes and lips
	Topical: Adapalene Tretinoin Tazarotene			Cheilitis and dry eyes Depression and psychiatric disorders Risk of pancreatitis with hyperlipidaemia	
Steroids	Oral: Prednisolone, prednisone	Eczema Contact dermatitis	Regulate gene transcription and reduce inflammation and immune response	Hypothalamic-pituitary suppression Skin thinning Diabetes	Consider the need for supplemental steroids at induction Use adhesives with caution if skin thinning has occurred Systemic absorption of topical steroids should be considered if potent steroids are used for a prolonged period of time over a large body surface area Measure serum glucose and electrolytes
	Topical: Hydrocortisone (mild) Fluticasone (potent)	Psoriasis Pemphigus		Osteoporosis Muscle wasting Peptic ulceration Hypertension Sodium retention, potassium loss	

Table 2. Medications for skin diseases with implications for anaesthesia

can increase the risk of gastric regurgitation and pulmonary aspiration. Children with reflux should be given prophylactic antacid medication before induction.^{6,7}

Physical Exam

Document the areas of skin affected and the site and size of any lesions. Consider potential sites for intravenous access and the ease of securing cannulae. Assess for signs of dehydration and malnutrition or ocular injuries, including the presence of ectropion. Children with EB may have oral-mucosal involvement leading to microstomia, and pharyngeal and laryngeal involvement is also possible. Assess the airway for xerostomia or microstomia with limited mouth opening. There may be limited neck movement due to hyperkeratosis in ichthyosis.

Laboratory Investigations

Children with severe disease or systemic treatment are at risk of anaemia from chronic disease or malnutrition.⁵ Measure a full blood count and serum electrolytes if significant fluctuations are expected perioperatively. See Table 2 for suggested investigations due to medications.

Other Considerations

A good rapport with the child helps facilitate smooth induction. Skin disease can have negative implications on a patient's mental health,¹² to which the anaesthetist should be sensitive. In children with very fragile skin or EB, consider sedative premedication to decrease the chance of friction or shear pressure on the skin at induction.⁵ Oral or intranasal premedication is preferable to intramuscular to prevent skin trauma and bulla formation at the injection site. Consider if topical local anaesthetic cream is appropriate. If used in children with EB, it should not be secured with an adhesive dressing; clingfilm is preferable.⁷ Consider a preoperative review by a dermatologist for all patients with severe or complicated skin disease to optimise their treatment before anaesthesia.

INTRAOPERATIVE CONSIDERATIONS

There is no contraindication to general anaesthesia with volatile or total intravenous anaesthesia or regional or neuraxial anaesthesia. Communication with the theatre team is essential to avoid unnecessary movement and trauma to the patient's skin, and careful positioning must be maintained at all times.

IV Access

Venous access may be challenging due to fragile vessels (EB) or thickened skin. Using ultrasound may improve success. Manually squeezing the limb over gauze is recommended rather than a tourniquet to minimise trauma.^{6,7} Securing intravenous cannulae may be difficult. They can be held in place with gauze, or a circumferential nonadhesive dressing may be required to facilitate security (Figure 1). Silicone tape ("Siltape") has also been used to secure intravenous lines and endotracheal tubes.

Monitoring

Intraoperative physiological monitoring (ECG, noninvasive blood pressure, and oxygen saturation) is considered essential. Clingfilm and thin gauze padding under the blood pressure cuff provides cushioning and helps reduce the likelihood of trauma.⁷

Invasive arterial pressure monitoring may be preferable for longer cases. The insertion site should be visible or regularly checked in case it is suboptimally secured and falls out.

For pulse oximetry monitoring, clingfilm can be wrapped around the digit, and the probe can be placed over it to measure oxygen saturation without causing skin trauma, as bullae may form from minimal friction.

Emollient use or the presence of skin scales may result in nonadherence of ECG electrodes. Extra contact gel can be used to maximise skin contact, and electrodes can be secured with gauze as an alternative to adhesives to avoid chemical skin damage (Figure 2). In children with EB, the adhesive material on electrodes should not touch the skin. A silicone film dressing can be placed under the ECG electrodes, and silicone tape can be used to secure them in place.⁷ Alternatively, needle electrodes can be used.

Airway

Lubricating the patient's lips and the facemask can prevent skin trauma from mask ventilation in children with EB. Gauze may be placed on the face underneath the mask to reduce pressure on the skin. The anaesthetist's gloves should be lubricated to avoid shear forces on the skin when holding the airway. However, this could make the airway more difficult to manage as it will be slippery. It is also good practice to lubricate any instrument that will enter the airway.



Figure 1. Use of gauze wrapped circumferentially around a limb to secure an intravenous cannula without the need for an adhesive dressing. (Photographs supplied by Dr C. Riley, Consultant Paediatric Anaesthetist, Sheffield Children's Hospital, Sheffield, UK).

When clearing oropharyngeal secretions, use a soft suction catheter and low-pressure suction to prevent mucosal trauma. Avoid oropharyngeal airways and touching the oral mucosa at all, if possible, as this could lead to life-threatening bullae formation.

Intubation

Difficult laryngoscopy should be anticipated, especially in children with ichthyosis, due to reduced mouth opening, limited neck movement, stomal ulcers and strictures, or poor dentition. A difficult airway plan should be made, and equipment should be immediately available. A smaller endotracheal tube (0.5 to 1.0 mm internal diameter smaller) may be necessary to reduce potential tracheal damage.^{5,6} There is a risk of mucosal damage or tearing from intubation or skin shearing if the endotracheal tube or intravenous lines are taped to the skin. Tying the tube in place may be a better alternative, for example, with saline-soaked ribbon with gel-lubricated gauze underneath the tie to provide padding (Figure 3). A supraglottic airway, not taped but constantly monitored, is an alternative. Consider placing the supraglottic device under direct vision using a laryngoscope rather than pushing it in. Bullae may form or friable tissues may bleed from shear forces, so the device should be generously lubricated, and a small size should be chosen if possible.⁶

Fibre optic intubation should be considered, particularly for children with EB, which avoids the potential trauma of laryngoscopy; the nasal mucosa is less vulnerable to bullae,⁶ and a nasal tube is easier to secure than an oral tube.

Suxamethonium is not contraindicated, but there is a potential risk of hyperkalaemia due to disuse atrophy, and skin damage could be caused by fasciculations.⁶ Deep muscle relaxation is recommended to aid smooth intubation.

Deep extubation may be considered, barring aspiration or airway concerns, as it potentially decreases the risk of coughing or airway trauma at extubation.

Fluid Balance

There is potential for the loss of large volumes of fluid from impaired skin integrity. Fluid balance should be closely monitored, and an arterial line and central venous access may be useful for major surgery and longer cases. Placing a urinary catheter has the potential to cause mucosal damage and strictures. It would seem prudent to avoid this if possible.

Thermoregulation

To avoid inadvertent intraoperative hypothermia, the ambient temperature in the theatre should be $>28^{\circ}\text{C}$. Oesophageal temperature probes may cause mucosal trauma, and a rectal or noncentral but less invasive alternative such as a tympanic thermometer may be preferable.⁶

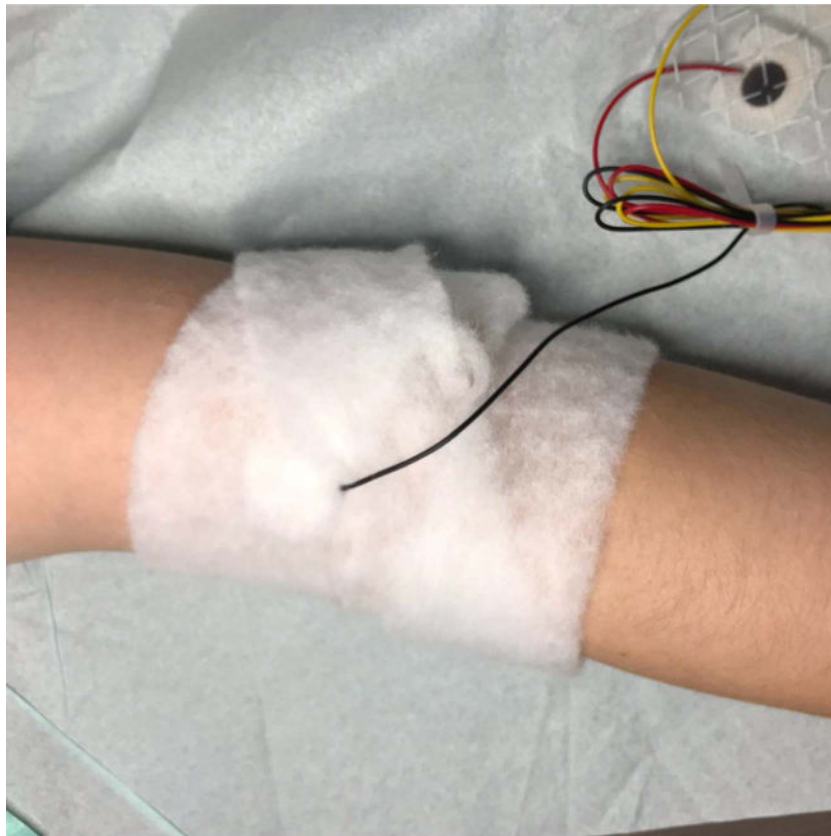


Figure 2. Use of gauze padding to secure electrocardiograph electrodes. Extra contact gel may be used under the electrode to maximise skin contact, and the electrodes can be secured with gauze as an alternative to adhesives to avoid chemical skin damage.

Infection Control

Postoperative sepsis is a concern due to impaired skin integrity. Strict asepsis and infection control measures should be followed.

Positioning

There is an increased risk of pressure sores as well as joint and nerve damage due to restricted movement of the limbs and neck. Care should be taken when transferring and positioning the patient with judicious padding of pressure areas. The epidermis can be prone to cracking in many skin diseases, so delicate handling should be used to prevent this, which may increase the risk of infection. Avoid using a slide board to transfer as shearing forces on the skin can cause large bullae, especially in children with EB. A nonadhesive silicone-based dressing can protect any blistered areas from further trauma.

If a tourniquet is required for surgery, skin padding underneath the tourniquet is essential. Ensure that the surgical skin preparation fluid cannot get under the tourniquet, as this can cause chemical irritation, burns, and pain.

Eye Care

Take care to prevent corneal abrasion due to ectropion, particularly if the patient is in the prone position. Eye lubricant may be used with silicone tape to protect the eyes in preference to sticky tape.⁷

Analgesia

Multimodal analgesia is recommended. Regional anaesthesia is not contraindicated, but exercise caution with skin preparation fluid such as chlorhexidine. Avoid harsh scrubbing by blotting the skin gently or spraying the skin with the antiseptic solution.⁵ Avoid inserting the needle over an affected area or active infection site. In children with EB, it is prudent to assess the skin condition for scarring from previous bullae or infection as haptic feedback and landmarks may be altered. The risk of ongoing sepsis should be considered before placing an indwelling neural catheter. If an epidural catheter is placed, gauze around the patient's waist can be used to help secure it instead of an adhesive dressing.



Figure 3. The endotracheal tube is tied into position using a soaked ribbon. Lubricated gauze underneath the tie provides padding to protect the underlying skin from pressure or shear forces.

POSTOPERATIVE CONSIDERATIONS

Assess the patient's temperature frequently and continue active warming in the postoperative period if necessary. Strict asepsis and infection control measures should be followed for wound care.

The child should be monitored in a specialist environment with the aim being early resumption of oral retinoids and emollients.

SUMMARY

Dermatological disorders in childhood range from common and mild to life-threatening but rare. Skin disease can impact every stage of the anaesthetic and should be evaluated in the preoperative assessment. The anaesthetist must balance prevention of further skin trauma or infection with the safe provision of anaesthesia. Elective surgery should be done in a specialist centre with input from the multidisciplinary team.

REFERENCES

1. National Eczema Society. Information and advice. Accessed March 31, 2023. <https://eczema.org/information-and-advice/>
2. National Eczema Association. Conditions related to eczema. Accessed March 31, 2023. <https://nationaleczema.org/eczema/related-conditions/>
3. Tjalma WAA. Burning of an ulcerated breast cancer during MRI: a lesson to be learned. *J Belgian Soc Radiol.* 2014;97(2):125.
4. Thakur BK, Murali MR. EMLA cream-induced allergic contact dermatitis: a role for prilocaine as immunogen. *J Allergy Clin Immunol.* 1995;95(3):776-778.
5. Bowen L, Burtonwood MT. Anaesthetic management of children with epidermolysis bullosa. *BJA Educ.* 2018;18(2):41-45.
6. Mittal B, Goodnough C, Bushell E, Turkmani-Bazzi S, Shappard K. Anesthetic management of adults with epidermolysis bullosa. *Aneth Analg.* 2022;134(1):90-101.
7. Birmingham Woman's and Children's NHS Foundation Trust. Care of child or young person with epidermolysis bullosa in theatre. Accessed March 31, 2023. <https://bwc.nhs.uk/download.cfm?doc=docm93jjm4n2799.pdf&ver=4387>
8. Hatch D, Sumner E. *Textbook of Paediatric Anaesthesia.* 3rd ed. Hodder Arnold; 2008.
9. DermNet. Harlequin ichthyosis. Accessed March 31, 2023. <https://dermnetnz.org/topics/harlequin-ichthyosis>
10. DermNet. Collodion baby. Accessed March 31, 2023. <https://dermnetnz.org/topics/collodion-baby>

11. British Medical Association and Pharmaceutical Society of Great Britain. *British National Formulary for Children Edition 85 (March–September 2023)*. BNF Publications; 2023.
12. DermNet. Psychosocial factors in dermatology. Accessed April 4, 2023. <https://dermnetnz.org/topics/psychosocial-factors-in-dermatology>



This work by WFSA is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view this license, visit <https://creativecommons.org/licenses/by-nc-nd/4.0/>

WFSA Disclaimer

The material and content provided has been set out in good faith for information and educational purposes only and is not intended as a substitute for the active involvement and judgement of appropriate professional medical and technical personnel. Neither we, the authors, nor other parties involved in its production make any representations or give any warranties with respect to its accuracy, applicability, or completeness nor is any responsibility accepted for any adverse effects arising as a result of your reading or viewing this material and content. Any and all liability directly or indirectly arising from the use of this material and content is disclaimed without reservation.